PHILADELPHIA NEUROLOGICAL SOCIETY.

March 27, 1899.

The President, Dr. James Hendrie Lloyd, in the chair.

Dr. Lloyd presented a young woman who had almost total internal and external ophthalmoplegia.

Dr. B. Sachs, of New York, remarked that instances of this complete form of ophthalmoplegia were, on the whole, rare. He had not seen a dozen cases in the course of many years. He thought that the diagnosis as made by Dr. Lloyd was entirely correct and that the case was of nuclear origin. Whether or not Hutchinson was right in believing that these cases were due to syphilis, is difficult to determine. He would not be so much inclined to ascribe them to syphilitic infection as he would some of the cases of partial ophthalmoplegia that are supposed to be nuclear.

A number of years ago Thomsen, of Berlin, described an unusual case in which there was specific infiltration of the oculo-motor roots, choking some fibers without destroying them, and giving rise to partial oculo-motor symptoms. That case had always stood out in his mind as an unusual example of root disease in contradistinction to what, on clinical grounds, would be supposed to be nuclear disease.

This case of Dr. Lloyd he was inclined to rank with those described by Wernicke under the head of polioencephalitis superior. In the majority of these cases, particularly in the acute form, there was a marked alcoholic history. Cases have been reported by the speaker and others, in which ocular symptoms were associated with poliomyelitis anterior, and it is evident that the affection may at times attack the gray matter in the floor of the third and fourth ventricles, or at other times involve the gray matter of the cord.

Dr. William G. Spiller said that although it was possible that the nuclei of the sixth nerves in the case were involved in the primary lesion, he would be more inclined to attribute the disturbance of function of the external recti muscles to impairment of the connection between the ocular centers by means of the posterior longitudinal bundles. The absence of any involvement of the facial nerves seemed to indicate that the sixth nuclei were not primarily diseased.

The case of Thomsen alluded to by Dr. Sachs resembled somewhat the one reported by Gibson and Turner (*Edinburgh Med. Journal*, 1897); in this a hemorrhagic infiltration was found in the oculo-motor nerve. A hemorrhage in the opposite frontal lobe was at first supposed to be the cause of the oculo-motor paralysis.

Dr. Spiller also referred to a case of hysterical ophthalmoplegia reported by Dr. Sachs.

Dr. James Hendrie Lloyd thought that the point made by Dr. Sachs in regard to root involvement was of interest. The possibility of this involvement could be excluded in the present case, because the third nerve was not exclusively involved. The fourth and sixth nerves were also paralyzed. In the absence of other lesions, the case could only be explained as one of those selective morbid processes involving the nuclei.

ERYTHROMELALGIA AND ALLIED DISORDERS.

Dr. B. Sachs, of New York, read this paper by invitation.

While rare diseases are not necessarily interesting, the reader referred to the importance of erythromelalgia, because of the discussion that necessarily involved the consideration of other disorders to which it is more or less closely allied. In his studies on the subject he attempted to answer the following questions: Does erythromelalgia constitute a disease *sui generis*, or is it a mere complex of symptoms which may be associated with a number of other diseases? Is it of peripheral or central origin? Is it always due to the same cause, or is it in any sense a purely nervous disease?

A brief review of the subject showed that the majority of writers who judge the matter from a purely clinical standpoint thought that the symptom-group of erythromelalgia was due to involvement of the posterior and lateral gray matter of the cord. This inference was based solely upon the association of erythromelalgia with such diseases as syringomyelia, Morvan's disease and tabes dorsalis. But one autopsy had been performed up to the present time on a patient suffering from erythromelalgia. In this case changes were found by Auerbach and Edinger in the posterior roots, but it is doubtful whether this disease of the posterior roots was at all responsible for the condition of erythromelalgia, while it may have had an important bearing upon the complicating tabes dorsalis. Examinations of sections of the peripheral nerves and of the arteries had been made by Mitchell and Spiller from an amputated toe in a typical case. Their findings seemed to bear out Mitchell's contention that while the disease may possibly be of central origin, it could also be explained on the assumption of a nerve-end neuritis.

In the patient detailed by Dr. Sachs the disease was typical for many months, until a large gangrenous ulcer developed on the dorsum of the foot. On this account amputation of the thigh, immediately above the knee, had to be performed, and opportunity was given for ample examination of the nerve strands and arteries in the leg. The histological examination which was made in conjunction with Dr. Alfred Wiener, of New York, proved that all the arteries of the leg, but particularly the popliteal and the anterior tibial, were markedly diseased. The lumen in the larger branches, as well as in the smaller, being almost entirely occluded. In the various nerves that were examined there was slight degeneration, altogether out of proportion, however, to the disease of the blood-vessels.

It is safe to assume that this obliterating endarteritis was present in the largest as well as the smallest blood-vessels of the thigh and leg. As the speaker's case was entirely uncomplicated, the findings prove that the symptom-group could be explained entirely on the assumption of peripheral and arterial disease, followed by nerve degeneration. He was thus led to agree with Mitchell and Spiller as to the probable peripheral origin of erythromelalgia, laying greater stress, however, upon the disease of the blood-vessels than upon the disease of the peripheral nerves; and while it is barely possible that in some instances the same or similar group might be caused by disease of the gray matter of the cord, that proposition was still to be proved. Everything pointed, however, to the conclusion that erythromelalgia did not represent a *morbus sui generis*, but was a symptom-group which might exist independently of all others, but might at times be associated with other diseases.

The speaker finally called attention to the fact that there were conditions following upon chronic heart and arterial diseases which resembled erythromelalgia very closely, and that from the point of view of morbid anatomy, there was some resemblance between intermittent claudication and the disease under discussion, only in the former the obliterating endarteritis, as shown by Erb, leads to temporary motor paralysis, while in erythromelalgia the obliterating endarteritis causes a series of sensory and vaso-motor disturbances.

Dr. Wharton Sinkler remarked that the paper recalled to his mind certain cases that he had seen resembling erythromelalgia without pain. The symptoms were more those of arterial disease than of nerve disease. In some cases there was intense flushing of one foot when the limb was pendent. This led him to think that the theory of Dr. Sachs was certainly tenable; but the conclusion cannot be escaped that in typical cases of erythromelalgia, as described by Dr. Mitchell, there is primarily disorder of the peripheral nerves, as shown by pain, and that coincident with the pain, there are disorders of circulation which are due to the nervous disturbance.

The president asked if it might not be of value to institute bacteriological examinations in these cases. The extreme involvement of the intima of the arterial system indicated the possibility of some microbic infection. The fact that the condition was limited to the vessels of one limb would of course be an objection to this view. This would also militate against the idea of a central origin. The possibility of thrombosis in the main artery of supply should also be borne in mind.

Dr. W. G. Spiller thought that the presence of gangrene was not sufficient to exclude a diagnosis of erythromelalgia. If further examination shows that the arteries are usually affected in this disease we may expect to find gangrene occasionally. It seemed strange, however, that in the case reported by Dr. Sachs, as well as in the one reported by Dr. Mitchell and himself, the disease was asymmetrical. If arteritis were the only cause, or the essential cause, we would expect to find the limbs bilaterally involved in erythromelalgia. Dr. Sachs had stated that the arteries in his case were more diseased in portions remote from the area of gangrene; that would seem to indicate that the disease was hardly of microbic origin, so far as the gangrenous area was concerned.

Dr. Spiller said that the atrophy of the thigh muscles and the exaggeration of the knee-jerk, mentioned by Dr. Sachs, were interesting features of the case. Exaggerated knee-jerk was also observed in the case reported by Dr. Mitchell and himself. The atrophy of the thigh muscles seemed to indicate that the anterior crural nerve was involved, and nevertheless the knee-jerk was exaggerated.

Dr. Spiller thought that Dr. Sachs had been very fortunate in saving his patient after so serious an operation, for surgical intervention in these cases has sometimes been fatal. In one case mentioned by Dr. Mitchell the patient died on the operating table, and in the patient reported by Dr. Mitchell and himself an ugly slough formed after the amputation of the toe.

The endarteritis described by Dr. Sachs agreed with the findings of Mitchell and Spiller, but Dr. Spiller was unwilling to assert from the findings of one case that the essential lesion of erythromelalgia was arteriosclerosis. This view may be correct, but it is exceedingly difficult to distinguish the primary lesion when nerves and vessels are diseased, as disease of either may lead to involvement of the other, and the tissues most involved are not necessarily the first affected. Dr. Mitchell and he had been unable to say which was the primary lesion in their case. Even if the process were primarily vascular, involvement of the nerves seemed to be necessary for the complete clinical appearance of erythromelalgia. In the recent thorough work on intermittent claudication by Erb, to which Dr. Sachs had referred, the symptoms of erythromelalgia were distinguished from those of intermittent claudication. Intense arteriosclerosis often exists without the signs of erythromelalgia.

Within the past six months Dr. Spiller had seen three cases presenting symptoms resembling those of erythromelalgia. One was a pedler who carried heavy weights on her arms, and walked all day. She complained of intense pain in her arms and hands, and said that her hands at times "got as red as blood." She suffered also from pain in her feet. The second case was that of a woman who stood nearly all day. She complained of much pain in her heels, and these parts were somewhat red. The third patient was a butcher, who complained of considerable pain and redness of the feet, and he had no knee-jerk, but no other sign of tabes was discovered.

The question had occurred to Dr. Spiller, Why do we not find symptoms of erythromelalgia in every case of neuritis? Possibly the disease is found only when the vaso-motor nerves are affected. It is not improbable that in different forms of neuritis different nerve fibers are diseased. We know that lead, alcohol and the toxin of diphtheria not only select certain nerves, but may select certain fibers within these nerves. Dejerine has shown that neuritis is not uncommon in tabes, and the ataxia may be in part due to this neuritis. It is not unreasonable to believe, therefore, that in some forms of neuritis the vaso-motor nerves are especially involved.

Dr. H. A. Hare said that he had not heard all of the paper, and did not know whether the gangrene was of the moist or dry variety, or whether the veins were affected as well as the arteries. From the occlusion of the arteries he should expect to find dry gangrene, whereas, if the veins were diseased, the gangrene would be of the moist variety. This had been the case in endarteritis in typhoid fever.

Dr. Charles S. Potts referred to a case that he had seen a number of years ago at the University Hospital. The patient, a young man, had one toe crushed by a stone. Shortly afterwards he developed ascending neuritis, and after a certain length of time this was followed by typical erythromelalgia.

Dr. A. A. Eshner thought that it was not a remote possibility that in erythromelalgia we had primarily to do with some toxic process, not necessarily a poison from without, but more likely a poison generated within the body. The objection arising from its peculiar localization would have to be disposed of as Dr. Spiller had suggested. We do not know why it is that certain poisons have certain affinities. Many toxic substances expend a part of their activity upon the blood-vessels, and some also upon the nerves. In erythromelalgia there may be a degenerative process involving the blood-vessels and an inflammatory process involving the nerves, and through this vicious circle there results an aggravation of the symptoms attending either process. It seems not impossible that in this *combination* is to be found the explanation of the peculiar train of phenomena that we see in erythromelalgia—why with simple neuritis or with simple arteriosclerosis we do not have erythromelalgia, while with the association of neuritis *and* arteriosclerosis symptoms of erythromelalgia develop.

In addition to the cases referred to there was another that was under Dr. Mitchell's care, and in which operation produced a successful result. In this patient, a man of about twenty-five years of age, the symptoms followed a traumatism of the foot, induced by a falling stone. He was subjected to nerve stretching and nerve excision, and the symptoms at once improved, and ultimately disappeared.

Dr. Joseph Sailer referred to a case which he had recently observed and which was in the wards of Dr. J. Chalmers DaCosta at the St. Joseph Hospital. When the man stood the feet swelled and became of a bluish, dusky color. The pain was not excessive, but the swelling and pain were increased by walking. Pressure on the posterior tibial, the anterior tibial and the sciatic nerves caused intense pain. An interesting feature was a surgical complication which had occurred.

Dr. F. Savery Pearce called attention to the fact that sometimes cases of periostitis have been differentiated with difficulty from erythromelalgia. He referred to a case in which Dr. Mitchell was in doubt as to whether it was one of periostitis or erythromelalgia.

Dr. B. Sachs in concluding the discussion, said that no bacteriological examination had been made. The fact that the arteries were more diseased higher up than in the vicinity of the gangrene would be sufficient to disprove the microbic origin of the disease.

In regard to the slight difference between the view of Drs. Mitchell and Spiller and that expressed by himself, he wished to be understood in this particular; he did not at all deny the importance of neuritis as a possible factor in the disease. He sided entirely with Dr. Mitchell and Dr. Spiller as against the majority of German writers who have, without microscopical study, concluded that this was a central disease. He thought that symptoms of erythromelalgia may be associated with some central disease, but in the uncomplicated cases that have been reported it is not necessary to seek any central nervous disease—the peripheral changes are sufficient to account for the condition. He believed that erythromelalgia would be shown to be chiefly, or very largely, of arterial origin. The changes in the arteries are so much more pronounced than those in the nerves that he thought it was hardly possible to make the nerves responsible for these changes.

There was one point which he had neglected to mention. Several of those afflicted with erythromelalgia have been tailors, or those who sat in a crouched position or ran sewing-machines, and he thought that it might be better to go back to a simple mechanical explanation rather than to seek for a bacteriological origin. He was inclined to doubt the bacteriological or toxic causation of the disease which, after lasting for years, showed relatively slight changes.

In reply to Dr. Hare he said that the veins had been examined and found to show only very slight changes. In his case there was moist gangrene of the foot and also dry gangrene of the toe.